2. Page 16, line 20 to 30 and page 17 lines 1 to 15, change to read:

*NYX* encodes a 481 amino acid protein, herein called nyctalopin, which has sequence similarity with members of the superfamily of proteins containing tandem arrays of the leucine-rich repeat (LRR) motif [10,13]. Such proteins are known to function in protein-protein interactions, especially in matrix assembly, and therefore nyctalopin may possibly be mediating specific neural connections between cells in the retina. Moreover, the presence of the 24 amino acid consensus: x-x-I/V/L-x-x-x-x-F/P/L-x-x-L/P-x-x-L-x-x-L/I-x-L-x-x-N-x-I/L (where I,V,L,F,P and N are single letter amino acid codes and "x" represents any amino acid) in the core protein with cysteine clusters flanking the LRR domain (see Figure 3B) **[SEQ ID NO: 2]**, qualifies nyctalopin as a new member of the subfamily of small leucine-rich proteoglycans (SLRPs) [10]. From a homology comparison of nyctalopin with other SLRP proteins, it is evident that nyctalopin is a unique member of this subfamily and the LRR superfamily in general. Nyctalopin has five putative consensus sequences (N-X-(S/T)) necessary for substitution by N-linked oligosaccharides or keratan sulfate [14], three of these sequences lie within the LRR region. The $NH_2$-terminal end of nyctalopin is predicted [15] to contain a membrane signal peptide with a putative cleavage site between amino acid 23 and 24, AWA-VG (Figure 3). In addition, the carboxyl-terminal region of nyctalopin contains a GPI-anchor signal sequence, including the requisite GPI N-terminal signal sequence (amino acids 339 to 379), the C-terminal hydrophobic region (last 22 amino acids) and a potential cleavage site at amino acids 445-447 [16] (Fig. 3B) **[SEQ ID NO: 2]**. . The identification of these sites was accomplished at the website www.expasy.ch/tools, and is well known to those skilled in the art. Thus, *NYX* codes for a GPI-anchored proteoglycan with a putative membrane signal peptide. Without being limited to a theory, these results suggest that the clinical features of complete X-linked CSNB can be explained by the presence of a mutant nyctalopin (or entire absence of nyctalopin) causing the disruption of selected connections or interactions between retinal neurons, including those of the retinal ON-bipolar pathway, possibly during early stages of embryonic development.

3. Page 22 lines 28 to 30 and page 23 lines 1 to 10, change to read:

Fourteen different mutations have been identified in *NYX*, none of which are observed in chromosomes from normal individuals. In nyctalopin, there are 11 leucine-rich repeats, which

are all highly conserved with respect to the consensus sequence in SLRPs, and these are flanked by cysteine clusters (see Figure 3B) [SEQ ID NO: 2] [10]. The deletion of a portion of the cysteine cluster in the amino-terminal portion of nyctalopin appears to be responsible for complete X-linked CSNB in six families, which highlights the importance of this conserved region. The mutation that causes a stop codon on the carboxyl-terminal side of the leucine-rich repeats and another cysteine cluster, likely affects the ability of the protein to anchor in the membrane, as the protein portion on the carboxyl-terminal side of this mutation is presumed to be important for GPI anchoring nyctalopin in the cellular membrane. Mutations that replace a consensus amino acid with another amino acid are presumed to disrupt an essential amino acid function. Mutations that result in the insertion ( or deletion) of amino acids in the protein are presumed to alter the folding of the protein

4. Page 30, lines 6 to 12, change to read:

Six other families were found to have an in-phase 24-nt deletion that results in the loss of eight amino acids - RACPAACA (see Figure 5B). Six of these amino acids form part of a conserved cysteine-cluster on the amino-terminal side of the leucine-rich repeats, as shown in Figure 3B [SEQ ID NO: 2]. Haplotype analysis of X chromosomes with this deletion mutation from each of the six families revealed nearly identical haplotypes, suggesting that these families share a common founder mutation. In three families, insertion mutations representing duplications of adjacent protein sequence add either six or three amino acids (Figure 3B) [SEQ ID NO: 2].

## AMENDMENTS TO THE CLAIMS

1. Please amend claims to read as follows

Claims 1-24 (Cancelled)

Claim 25 (Previously Added) An isolated or recombinant DNA molecule encoding the amino acid sequence of SEQ ID NO: 2.

Claim 26 (Previously Added) The DNA molecule of claim 25 comprising a nucleotide sequence corresponding to SEQ ID NO: 1.